MYOCLONUS-EPILEPSY WITH A REPORT OF TWO ADDITIONAL CASES.*

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This condition is one of the numerous forms of the epilepsies of which there has been but a small number of cases reported.

A review of 2150 cases of epilepsy admitted to the Craig Colony for Epileptics, since its opening in February 1896, shows but seven cases of myoclonus-epilepsy. The first of these was reported by L. Pierce Clark in *Archives of Neurology and Psychiatry,* Vol. II, 1899. Three of these were reported by L. Pierce Clark and T. P. Prout in their excellent paper in the *Journal of Insanity* in 1902; the fifth by L. Pierce Clark and myself, the last two being now reported for the first time.

The first case on record of paramyoclonus multiplex reported was that by Friedreich in 1881. A search through the earlier literature shows histories of cases which were undoubtedly the same condition. He describes a patient, aged 50 years, who developed quick clonic contractions of symmetrical muscles of extremities. These contractions resembling those produced by an electric current, ceased during sleep; increased by peripheral mechanical stimuli. They were diminished or entirely inhibited by voluntary movement. Muscles of trunk and face escaped. Nutrition and mechanical and electrical irritability unaltered. Increased knee jerks, no affection of sensation. No interference with motor power or coordination of involved muscles.

His case was a result of a severe shock, and the spasms, after lasting several years, rapidly ceased, recurring later and continued until death.

Many variations from this description have been seen in cases reported later. In some there is considerable locomotor effect, in others little or none.

Face and trunk muscles are frequently involved. Voluntary movements often increase the myoclonic movements instead of diminishing them. Knee jerks may be normal or diminished instead of increased.

It attacks groups of muscles which cannot be thrown into

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*Read before the Rochester Pathological Society, May 2, 1907.

contraction voluntarily and movements show no coordination as occurs in movements produced at will. In the majority of cases the proximal segments of the limbs are most affected.

Mental excitement and physical fatigue often provoke the movements. Excess of indican has been present in some cases when the myoclonic movements were most marked. In other cases it was absent. The movements are of a type that cannot be produced voluntarily.

Some of the phenomena of hysteria may be present in myoclonus epilepsy as in ordinary epilepsy.

Clark in his article in *The American Journal of Insanity* in 1902 has compiled a list of 57 cases of myoclonus epilepsy reported up to that time. Since then several cases have been reported in the literature. The most recent being eight cases reported from the Colony for Epileptics at Bielefeld in a population of 2000 epileptics.

Aldren Turner in his recent work on Epilepsy reports but two cases of myoclonus epilepsy in one thousand cases of epilepsy that came under his observation.

Many of these cases have been confused with motor aura and incomplete seizures.

Unverricht in 1891 in his monograph, "Die Myoclonie," describes several cases of a family type.

Etiology.—The same causes which predispose to epilepsy, insanity, imbecility, etc., are found in these cases, viz., alcoholism, insanity, tuberculosis, epilepsy and other signs of family degeneracy. These patients themselves show many signs of degeneration.

The exciting cause, if one is given, is frequently one which has little or no bearing on the condition in question. In one of the two cases reported in this paper no cause is assigned except that patient was always "nervous." In the other jerkings appeared a short time after patient had measles.

Sex.—Males seem to predominate. In those at Bielefeld 5 were males, 3 were females. In the 57 collected by Clark in 1902 the ratio was 5 males, 3 females. In the seven at the Colony, 4 were males, 3 were females.

Most of the cases develop before adult life although some do occur later, one reported in this paper claiming that the myoclonic movements did not appear until she was over 30 years of age.

Turner's two cases were women and in them the myoclonus was present for many years before the first epileptic seizure.

Pathology.—Friedreich and Unverricht believed the disease to be due to excitation of the motor ganglia of the cord. Unverricht also considered an additional involvement of the cortical motor ganglia. Others have described the condition to lesions in the medulla oblongata, cerebellum, corpus striatum, optic thalamus and cerebral cortex. As has been frequently stated, it would seem if the lesion was in the spinal ganglia we would have trophic changes in the muscles.

Clark and Prout conclude that the lesions appear to be in the cerebral cortex involving the nucleus and the intra-nuclear network of cells of both sensory and motor types. Its pathogenesis appears to be an intoxication or an auto-intoxication of these cortical cells, probably brought about by a faulty chemotaxis of these same cells because of an inherent organic anomaly.

Clark reports a case from the Craig Colony in which an autopsy was made and a microscopical examination of the brain showed the following condition: General chromatolysis, absence of nuclear membrane and granular (often swollen) nucleus in every portion examined (Sections taken from both motor and frontal areas). He lays stress on the fact that the large pyramidal cells of the third layer are especially involved.

J. Ramsey Hunt believes that there is a coexisting degenerative affection of the cerebro-spinal axis, the epilepsy referable to the upper, the myoclonus to the lower centres. He found a marked hypertrophy of the muscle fibers in a case of myoclonus multiplex.

J. Risien Russell's hypothesis is that according to the phenomena, the motor neurones of both the brain and spinal cord may be affected, and that the neuroclonic state existing in the neurones of the anterior horns of the spinal cord or their homologues in the medulla oblongata and pons, may give rise to myoclonic spasms, whereas an altered state in the neurones of the cerebral cortex may be subsequently and secondarily induced, giving rise to epileptic seizures. Or again the altered condition of the cerebral cortex, and the consequent epilepsy, may be primary, and the neuroclonic state of the neurones of the spinal cord may appear subsequently.

This hypothesis of Russell's appears to be very reasonable when the symptoms are given careful consideration.

Diagnosis.—This condition is confused at times with pseudo-myoclonic contractions (which the patients themselves characterize as "jerks"): isolated tics of epilepsy; post-hemiplegic choreic movements associated with epilepsy; choreic epilepsy; hysteria; and myoclonia in general paresis.

The characteristic grand mal seizure and movements which could not be produced voluntarily should be sufficient to exclude hysteria.

The pseudo-myoclonic movements, "jerks" are bilateral flexure movements of the upper extremities although cases are seen in which these premonitory "jerks" involve the lower extremities. Some are on the border line between epilepsy and myoclonus epilepsy.

Various tics in epilepsy are confined to the face; are not coordinate and usually voluntary or reflex emotional acts, acts whose general character continues unchanged although exaggerated by emotions.

One case admitted to the Craig Colony showed multiple tics involving especially face and right arm. These movements were constantly present to a greater or lesser degree but increased markedly 24 to 28 hours before seizures. Patient had a marked neuropathic family history.

In choreic paresis and in infantile spasmodic hemiplegia (without palsy) the convulsions begin and involve most frequently parts once paralzed. The choreiform movements are rhythmical and unilateral. Unilateral atrophy also occurs.

In general paresis the mental state, speech, pupils, etc., should make the diagnosis clear.

The diagnosis is made on the presence in an epileptic of bilateral, electrical shock-like convulsions occurring in paroxysms and involving no impairment of consciousness and at intervals the occurrence of grand mal seizures, epileptic in nature.

It may appear as a pre-epileptic or post-epileptic phenomenon or as a condition independent of the distinct epileptic seizure. In the latter case it is sometimes considered as an equivalent of the seizure.

Prognosis—Periods of freedom from myoclonic movements of variable length occur but there is no permanent cessation. The mental condition may change but little. Death may occur in status myoclonus or from intercurrent disease as pneumonia.

Treatment—This must be along general lines tending to place a patient in the best possible physical condition.

Chloral hydrate and amylene hydrate have a marked effect in controlling myoclonic movements. Bromides have an excellent effect in some cases.

Report of two new cases follows:

E. C. B., age 39 years. Single. Common school education.

Family History—Father living, aged 71 years. He is an alcoholic and has been frequently intoxicated during the past twenty-four years. Said to be a worthless character. No definite history of syphilis can be obtained but as he was an old alcoholic soldier and his wife had several miscarriages it would seem probable that he had syphilis.

Mother was a nervous woman who had frequent headaches, fainting spells and attacks of vertigo, but no fits or movements similar to those patient has. She died at age of 39 years. Assigned cause of death was confinement and Bright's disease.

Maternal grandmother died at age of 87 years. She enjoyed good health until late in life, when she developed some nervous trouble.

Patient eldest in family of five children. One brother died at 19 years from pulmonary tuberculosis. One sister was sickly and died at age of two years from some intestinal trouble. Two sisters married, both of whom are in poor health, exact condition unknown. One has no children, the other has had several who have been sickly and died young. She has one daughter living and well. One of the children who died, a son, had convulsions during his last illness at five years of age. He is said to have been a bright boy but frequently cried without apparent cause.

Two maternal aunts are troubled with some gastro-intestinal disorder, the exact nature of which cannot be ascertained.

Personal History—Patient was born at full term. Nursed by mother. Nothing can be ascertained as to delivery or early infancy of patient. No history of any traumatism. No history of difficulty in learning to walk. Had measles at six years. She is said to have been ill for three months with scarlet fever at age of nine years. During the period of this illness she had convulsions and was totally blind for an unknown period.

Patient claims that her first seizure, resembling a fainting spell, occurred when she was eighteen years of age. She was at that time working as a clerk in a store and had to give up her position. The seizures recurred and became more severe until she had a general convulsion at age of twenty-two years. During this seizure she fell against radiator burning her right hand. Usually has no aura, but at times has a tingling in her fingers. Previous to her admission to the Colony she is said to have had seizures at intervals of from one week to two or three months. They occurred usually in series and were followed by a marked prostration and stupor. The first seizures in the series were severe and the later ones mild. Face and hands said to have been affected first. She had bitten lips and tongue during seizures, and received many injuries by falling at those times, viz., burns, fracture of nose, contusions about head and arms.

Patient complained of a pain and a "drawn feeling" in the head after seizures. At age of thirty-six she had a slight paralysis of left hand following seizures. This disappeared after two months. She has had as many as eight seizures in twenty-four hours. Is said to have been jealous, stubborn, depressed and at times hysterical.

Physical examination on admission to the Craig Colony, June 15, 1905. Brunette of spare build. Height 4 ft. 10½ in. Weight 71½ lbs. Has an anxious expression. Ears have adherent lobules. Small inferior maxilla. Palate has a narrow and moderately high arch. Moderate scoliosis. Slight facial asymmetry. Thyroid not enlarged. Skin and mucous membranes pale. No evidence of syphilis. Slight cyanosis of feet. Eyes—No ptosis, exophthalmus, nystagmus or strabismus. Twitching of eyelids. Cornea and conjuctiva are markedly sensitive. Vision—right eye, 20—40: left eye, 20—50.

Complains of an occasional tinnitus aurium. Gustatory sense normal. Olfactory sense impaired on both sides.

Chest negative. Has always been troubled with constipation. Appetite poor. Some pyrosis and distress of epigastrium after eating. Considerable prominence of abdomen in general, apparently due to accumulation of gas in intestines.

Breasts atrophied. At age of 27 years she had both ovaries removed as she was told this would cause her seizures to stop. Menstruation began at 14 years and was irregular up to time of operation. Complains of almost constant vertigo, suboccipital headache and pain in vertex. General feeling of weakness and exhaustion.

Tactile, pain, temperature, localization and stereognostic senses normal. Dermatographia absent. No anesthesia of ulnar nerve.

Slight abdominal and epigastric reflexes. Very active patellar jerks. Plantar is of lively flexion type. Active wrist and elbow jerks.

Patient right handed. Dynamometer shows no inequality between two sides. Handgrasp only moderately strong for her general physique.

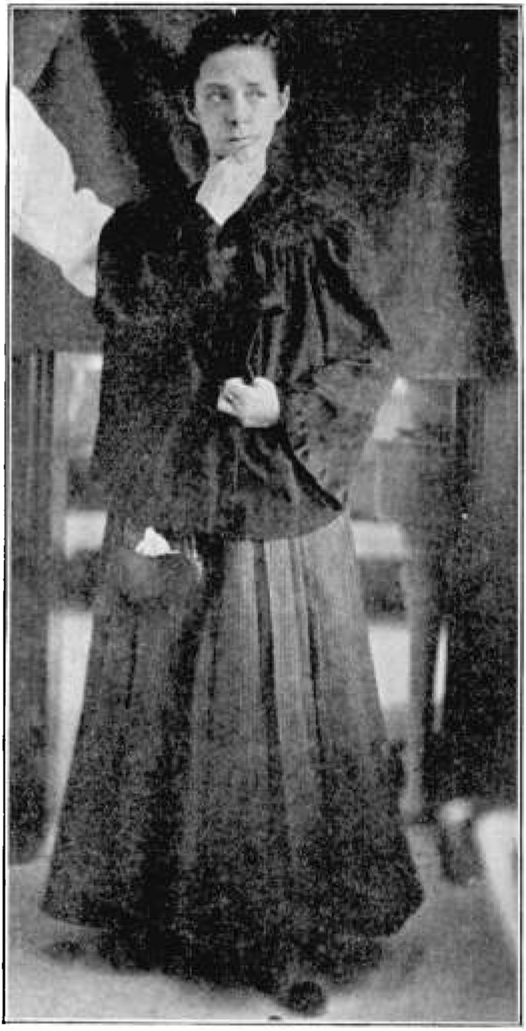

Fig. 1. E. C. B. Holding inferior maxilla with right hand and left hand grasping clothing tightly, in an effort to control myoclonic movements.

No signs of paralysis except that in showing her teeth muscles on right are more active than on left. General musculature is poor. Sense of position normal. Because of presence of myoclonic movements patient could not walk without assistance. Mentality fair. Speech interrupted but slightly by movements.

Shortly after her admission to the Craig Colony, she had a series of grand mal seizures which was followed by a state of mental confusion accompanied by illusions and hallucinations of sight and hearing. Disoriented. Very lachrymose and then would suddenly change and laugh in a meaningless way. She dressed herself and went to the table for her meals. At another time she had marked myoclonic movements following grand mal seizures. Seizures are usually grand mal in type and occur at intervals of several weeks.

Following is a brief description of a myoclonic period: Patient appears slightly agitated, is lachrymose. When walking with support there is a marked extension of right leg at knee from time to time. Very unstable equilibrium, especially when weight is borne on left leg. Bilateral clonic nodding movements, at times muscles on right side of neck drawing head to that side, occasionally similar movement of head to left. Smacking, tasting and munching movements of jaws. To and fro movements of tongue when mouth is open. Frequent marked clonic movements of muscles of mastication causing lower jaw to be depressed and then elevated. Patient holds inferior maxilla with her hand endeavoring to control movements. (See photograph.) Swallowing movements frequent. When mouth is closed there is a lifting of chin and compression of lips at frequent intervals.

Muscles of eyelids and forehead occasionally involved. Drank water without difficulty when glass was held to lips by nurse. No ejection of food from mouth. Active involvement of temporal muscles but only occasionally of occipito-frontalis. Complains of a "drawn sensation" in temporal and occipital regions.

When sitting or walking the muscles of back and back of neck are markedly involved in lightning-like jerks. No tremor of tongue. No nystagmus.

Can walk about room by balancing against walls, chairs, beds, etc., or having some assistance from nurse.

When lying on abdomen there is an occasional myoclonic movement of erector spinæ. Lying on back there is a marked myoclonic flexion of lower extremities, especially right. Right upper extremity, then left markedly involved. Flexors more than extensors.

Knee jerks exaggerated. Patellar clonus on right side. No ankle clonus or Babinski reflex. No increase in reflexes of upper extremities.

Abdomen prominent, walls are not rigid nor apparently involved in myoclonic movements. Peristaltic movements apparent in lower abdomen. No borborygmus.

Passive movements of extremities resisted. (Patient said she was endeavoring to exert some control over movement.)

No diaphragmatic grunt. No hiccough. Patient says she feels worn out and discouraged during myoclonic periods.

When not spoken to and allowed to lie quietly in bed, the movements are somewhat less frequent, but when spoken to become exaggerated.

She says she often has a sensation of tingling formication, followed by a feeling of coldness over entire body, but more in left arm and leg, especially former. Feels better when sitting up sewing, although when working at latter, she frequently sticks needles or scissors' points in fingers and has great difficulty in threading needle.

Urine examined during myoclonic periods shows no increase in indican. Sometimes urinates involuntarily during myoclonic periods. No impairment or loss of consciousness. Complains frequently of anorexia and insomnia.

Patient states that previous to her first seizure at age of eighteen years she was nervous, could not stand any excitement and had frequent crying spells. No myoclonic movements until she was about 34 years old. They first appeared in hands and feet, later in head. Movements have gradually become more severe and frequent. Movements are frequently more marked following seizures than preceding them. Marked vertigo accompanies periods.

She says the weather has no influence on the movements and this has been corroborated since her residence here. Electrical reaction normal.

This patient has not been entirely free from the movements at any time since her admission to the Colony.

E. J. W. Age 15 years. No occupation.

Patient is second child in family of three children (two boys and one girl, all of whom are living).

His brother, aged 11 years, has jerkings and has had "two or three fits."

Father died at 29 years from typhoid fever and is said to have had nervous prostration. Mother died at 23 years from tuberculosis. She had convulsions when a child. On his mother's side there has been a number of cousins who have had convulsions. Mother's people were "nervous." Father or mother was not intemperate. Maternal grandfather died at 40 years from tetanus. Otherwise grandparents negative.

Patient born at full term. Labor "quick and normal." Delivery natural. No injury during delivery. Weighed eight pounds at birth and was a strong baby. Had no spasms directly after birth nor was he paralyzed. Nursed by mother. No indigestion nor prolonged crying in infancy. Dentition began at four months and was not accompanied by convulsions. No pavor nocturnus. Began to walk without difficulty at age of fourteen months. Did not show any swollen glands or signs of rickets. No epistaxis. No injury in early life. Had pertussis at nine years and measles at ten; both ran ordinary course.

Patient says he started to school when he was ten years old, could not go earlier because he lived so far away from school.

First attack occurred when he was eleven years of age. He was driving a horse when it fell down and broke the harness and shafts. That night he had first convulsion just before going to bed. Supposed cause was fright. Second attack occurred three days after first. Attacks have occurred nearly every night during the last two years. Has had a few attacks in day time. Attacks are not as bad as formerly, but are more frequent. One attack is described, "He screeched and shook for about ten minutes." Feels badly in head after each attack. No aura. Vary in severity. Sometimes he says his right and sometimes his left side has a pain in it after a fit, more often the right. Patient is conscious in light attacks. No paralysis or aphasia following seizures. Has had seven attacks in 24 hours. Rather small eater. Patient says that for about a year preceding his fright he had what he called "jerkings." They began a short time after he had the measles.

Physical Examination—Rather slight. Hair brown and abundant. Poor musculature. Height, 3 ft. 9½ in. Weight, 74 lbs. Head well shaped. Palate high, some lordosis. Is flatfooted. Scapulæ stand far apart. No eruptions, scars or bruises. Skin and mucous membranes pale. No evidence of syphilis. Thyroid not enlarged. Eyes negative as also ears. Taste and smell normal. Chest somewhat flattened. Lungs and heart normal. Digestive organs normal.

Patient says he is not as strong as formerly. No vertigo except after seizures. Seldom has a headache. Following some attacks he has a pain on right side just below costal border. Tactile sense, slow in responding, seems diminished in all parts of body. Pain sense diminished. Temperature sense normal. Stereognostic sense poor. Reflexes negative. No evidence of any paralysis. Co-ordination and muscular sense normal. Has frequent jerkings of upper extremities, chiefly on right side, sometimes head jerked also. These movements occur frequently and are apparently more frequent when he closes his eyes. Slight tremor of tongue. Mental state fair.

November 1, 1905—Flexor muscles of leg are most often affected in convulsive movements. At times they are almost continuous changing from one group to another. No loss of consciousness. At times has great trouble in dressing and walking.

July 1, 1906—Continues to have much jerkings of his muscles, chiefly in arms, legs, head and lips. More on right than on left. They are worse in early morning, at which time he is quite helpless.

October 1, 1906—Movements are present much of the time, although some days he is entirely free. They are absent during sleep. All muscles seem involved with no distinct order of invasion. Sometimes upper extremities and sometimes lower. At times neck muscles. Always worse in morning.

January, 1907—Pale and anemic.

February 16, 1907—Myoclonic movements continue. At frequent intervals, especially in morning, although at times they are present during entire day. Recently received a severe contusion of tissues about left eye by falling during a seizure. Myoclonic movements involve trunk extremities and facial muscles. To-day, directly following a grand mal seizure, he had loss of plantar, patellar, wrist and elbow reflexes. Wets bed every night. Ordered strontium bromide grs. x, sodium bromide grs. v, chloretone grs. iii, t.i.d.

February 27, 1907—Is in a semi-stuporous state. Occasional myoclonic movements on right side. Chloretone stopped.

March 13, 1907—Cold, rainy day. Myoclonic movements are very marked. (They were practically absent for past two weeks.) They are general and involve principally the flexors. Muscles of abdominal wall markedly involved. No nystagmus. Pupils widely dilated and react but slightly to light. Some spasm of eyelids but not much of facial muscles. No impairment of consciousness. Reflexes not exaggerated, except plantar which is toe flexion in type. No ankle clonus. Flexors of toes are first involved, then foot, then entire extremity on each side. No apparent involvement of muscles of back. Movements increased when talking.

March 15, 1907—Marked myoclonic movements during early morning and up to 10 a.m., when he had a grand mal seizure which was followed by marked perspiration and complete cessation of myoclonic spasms for several hours, after which they reappeared and were very severe, but again ceased when patient was given chloral hydrate grs. x, potass. bromide grs. xv. Developed difficulty in swallowing and seemed to have a spasm of laryngeal muscles producing difficulty in respiratory act. Occasional loud grunt.

March 16, 1907—Abdomen is now negative. Patient had no myoclonic movements for several hours after yesterday's grand mal seizure. They returned late in afternoon and became more severe.

He developed difficulty in swallowing and seemed to have inspiratory difficulty. Auscultation over chest, negative. Grunt heard occasionally as though respiratory muscles were involved in myoclonic movements.

March 28, 1907—Pupils well dilated. Wrist jerks exaggerated on both sides. No difficulty in swallowing fluids. Myoclonic movements involving sterno-cleido-mastoid, trapezius, deltoid and flexors of forearm and hand on both sides but not synchronously. Spasm of both eyelids is synchronous and bilateral.

No exaggeration of elbow jerks or knee jerks. No ankle clonus or Babinski reflex.

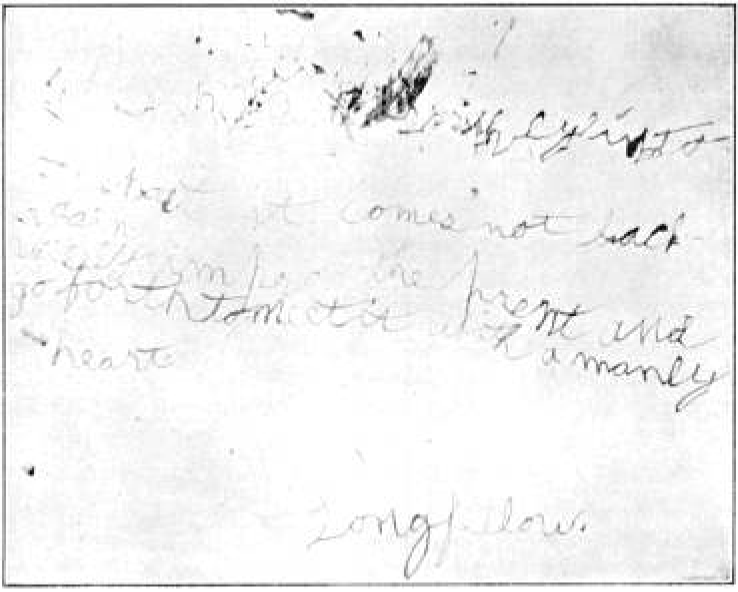

Fig. 2. Sample of writing of E. J. W., showing inability to control myoclonic movements.

Some borborygmus. No hippus.

March 29, 1907—Marked myoclonic movements to-day which involve muscles of back, abdominal wall, occipito-frontalis, right face, eyelids, flexors of lower extremities and all muscles of upper extremities. Right upper extremity affected much more than is left, arm being raised from side. Head turned toward left but not held rigidly.

Occasional grunt accompanies severe myoclonic movements. Facial muscles are not as markedly involved as are muscles of right arm. Some difficulty in speaking. Reflexes not exaggerated. Swallows without trouble.

Tapping muscles, excitement, etc., increases movements. Pupils normal.

General convulsion occurred about 10 a.m. Head turned markedly to left, pupils widely dilated, no cry before, but did occur after seizure (at time of cry patient appeared to have an atypical seizure as pupils dilated and there were some clonic movements of upper part of body). All parts of body involved. Saliva blown from mouth. Slight stertor and very brief period of cyanosis. Reflexes, tested immediately after cessation of clonic movements, were absent except for slight plantar flexion.

Flatus passed but no fecal matter or urine. Pupils returned to usual size directly after convulsive movements ceased. Moderate increase in pulse rate. Two minutes later reflexes were active but not exaggerated.

April 3, 1907—Myoclonic movements marked since 6 a.m. Mild seizure at 7.40 a.m. Thirty minutes after taking amylene hydrate by mouth all movements ceased and patient was able to sit up in bed and read.

April 5—Jerkings slight until 5 p.m., when they became very marked and he had a grand mal seizure at 5:15 p.m., after which he perspired very freely for several hours and was very restless. Myoclonic movements returned at 7 p.m.

April 6—Myoclonic movements continued very severe all night so that patient could not sleep.

April 9—Dark, rainy day. Marked myoclonic movements involving especially flexors of extremities both distal and proximal positions. Increased cremasteric reflexes. Diaphragmatic grunt. Slight lateral movement of eyeballs in some movements. Back muscles involved markedly also. Fibrillary twitching observed in posterior aspect of thighs. Consciousness unimpaired.

Seizures, both grand mal and abortive or atypical types, vary in frequency. Grand mal occurring from two to twenty-two times per month, the abortive form more frequently. Electrical reaction of muscles normal. At no time has there been an increase of indican in the urine.

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Excellent biographies are appended to articles of Clark, Hunt, Dana and Russell.